Ruptured Aneurysm of the Sinus of Valsalva into the Right Atrium without Ventricular Septal Defect: A Case Report and Literature Review

Shuo-Tsan Lee, Tsung-Hsien Lin, Ho-Ming Su, Chaw-Chi Chiu,[1] Wen-Chol Voon, Wen-Ter Lai, Sheng-Hsiung Sheu, and Young-Tso Lin[1]
Division of Cardiology, Department of Internal Medicine, and [1]Cardiovascular Surgery, Department of Surgery, Kaohsiung Medical University Chung-Ho Memorial Hospital, Kaohsiung Medical University, Kaohsiung, Taiwan.

Aneurysm of the sinus of Valsalva (ASV), frequently associated with ventricular septal defect (VSD), is a rare cardiac disease that may be acquired or congenital. Rupture of an ASV, rare in the noncoronary cusp, usually produces serious hemodynamic change and carries poor prognosis if not treated surgically. We present the case of a 55-year-old female who came to us complaining of exertional dyspnea. Transthoracic echocardiography and aortography showed a noncoronary cusp ASV with rupture into the right atrium but without VSD. Because of high left to right shunt flow, she underwent successful surgical intervention with aneurysm repair approached from both the aorta and right atrium with a knitted Dacron patch. This was a rare case of noncoronary cusp involvement in ASV that ruptured into the right atrium without VSD.

Key Words: aneurysm, sinus of Valsalva, rupture, right atrium, noncoronary cusp
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Aneurysm of the sinus of Valsalva (ASV) is a rare cardiac disease that may be acquired or congenital. Congenital ASVs are confined to one aortic sinus and consist of a localized diverticular outpouching of the coronary sinus. They may be associated with other congenital anomalies, the most common of which is ventricular septal defect (VSD). It is usually an asymptomatic condition, but when it ruptures or a fistula develops, symptoms occur and the condition deteriorates rapidly. We report a case of a 55-year-old female patient who suffered an ASV rupture into the right atrium.

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Address correspondence and reprint requests to: Dr. Tsung-Hsien Lin, Division of Cardiology, Department of Internal Medicine, Kaohsiung Medical University Chung-Ho Memorial Hospital, 100 Tzyou 1st Road, Kaohsiung 80708, Taiwan.
E-mail: lth@kmu.edu.tw

Case Presentation

A 55-year-old female patient had been diagnosed with an atrial septal defect with cardiac murmur 10 years before this presentation. She denied having diabetes mellitus or hypertension and reported no major trauma history. She visited our hospital because she became fatigued easily and had intermittent palpitation. She also had exertional dyspnea with intermittent and retrosternal chest tightness, characterized as an oppressive sensation that persisted for about 30 minutes. She denied any paroxysmal nocturnal dyspnea or orthopnea. No obvious toxic sign was found before admission.

Physical examination revealed an engorged jugular vein (6 cm above Lewis angle). A continuous heart murmur was heard over the left upper and lower sternal border. Moist rales were heard over bilateral basal lung fields. Mild pitting edema was found over both lower legs. Positive laboratory tests included microcystic anemia (hemoglobin

9.6 g/dL) and hyponatremia (Na 131 mmol/L). A 12-lead electrocardiogram (ECG) showed left ventricular hypertrophy and plain chest x-ray showed cardiomegaly and atherosclerosis in the aorta. Transthoracic echocardiography showed a suspected noncoronary cusp ASV with rupture into the right atrium (Figure 1). Transesophageal echocardiography was suggested, but the patient refused. She underwent cardiac catheterization, which showed oxygenation step-up in the right atrium, pulmonary hypertension, and left-to-right shunt from the aorta to the right atrium (Figure 2). The flow ratio between the pulmonary and systemic circulation (Qp/Qs) was 2:1. The clinical diagnosis was ruptured ASV into the right atrium. The patient underwent surgical intervention with repair of the ruptured aneurysm, approached from both the aorta and right atrium and using a knitted Dacron patch. After surgical intervention, she had an uneventful recovery and received regular follow-up at our clinic. Repeated echocardiography after surgery showed no residual shunt.

Discussion

ASVs arise immediately distal to the aortic valve and are named, more commonly, according to the coronary artery ostia that arise from the sinuses of Valsalva (right, left and noncoronary) [1]. The first description of aneurysmal enlargement of one of these sinuses with intracardiac rupture is attributed to Hope in 1839 [2]. Subsequently, Thurman published a series of six cases in 1840, including the one described by Hope, and noted the importance of the anatomic relationship of these aneurysms to the chambers of the heart [3].

ASVs are uncommon, being noted in seven of 8,138 autopsies and in 0.14–0.96% of open heart surgery cases [4–6]. Ruptured ASVs are most common in men (two-thirds of patients) and in the third or fourth decade of life (50–60% of patients). In 1919, Abbott clearly established a congenital etiology for ASVs [7]. The frequency of congenital ASVs is highest in the right coronary cusp, lower in the noncoronary cusp, and rare in the left coronary cusp [6]. The incidence of congenital ASVs appears to be higher in Asian populations, but the reason for this is not clear [5]. Acquired ASVs can be the result of several pathologic processes, including luetic degeneration, bacterial endocarditis, trauma, cystic medial necrosis, and atherosclerosis [4,8,9]. Acquired aneurysms are more evenly distributed among the three sinuses, with 44% arising from the right coronary sinus, 23% from the left coronary sinus, and 10% from multiple sinuses.

There is a frequent association (30–60%) between VSDs and congenital ASVs [10,11]. Without careful attention, these VSDs may remain undiagnosed, particularly if the wall of the aneurysm descends to occlude flow through the VSD [12]. Aortic valve abnormalities, primarily abnormalities that lead to aortic insufficiency, are common. The degree of aortic insufficiency and an assessment of its effect on left ventricular function are, therefore, important.

Most patients with unruptured ASVs are asymptomatic. However, unruptured aneurysms may produce symptoms when they become infected or impinge on other structures. Aneurysmal extension into the right ventricular outflow tract may produce obstruction, and extension into the ventricular septum can cause medically refractory ventricular tachycardia or affect the left ventricle [13].

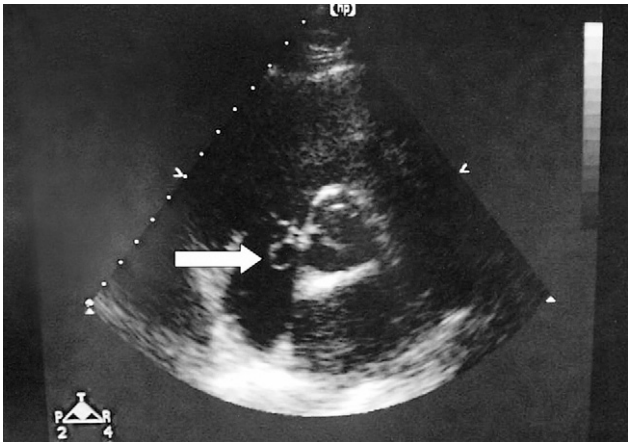

Figure 1. *Transthoracic echocardiography discloses an aneurysm of the sinus of Valsalva in the noncoronary cusp (arrow).*

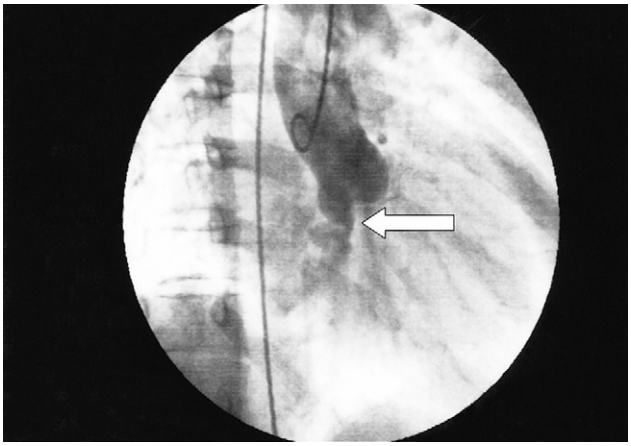

Figure 2. *Aortography shows left-to-right shunt from the aorta to the right atrium via a ruptured aneurysm of the sinus of Valsalva (arrow).*

In one-third of patients, left-to-right shunting immediately following the rupture of an ASV into the right side of the heart produces acute dyspnea and substernal chest pain that is often associated with epigastric or right upper quadrant abdominal pain. However, half of patients note the gradual onset of dyspnea, fatigue, chest pain, and peripheral edema over several months or even years following rupture of an ASV, and the remainder of patients are still asymptomatic at the time of diagnosis [10,11].

Venning, in 1951, was possibly the first to diagnose acute rupture of an ASV during life [14]. The diagnosis of ruptured ASV is generally made by history and physical examination in connection with echocardiography and angiography to define the precise anatomy of the aneurysm. As described by Morch and Greenwood, the murmur associated with the most commonly ruptured ASVs is a harsh continuous murmur. This must be distinguished from the murmur of a patent ductus arteriosus, VSD associated with aortic insufficiency, aortopulmonary window, coronary arteriovenous malformation, or pulmonary arteriovenous malformation [15]. A continuous murmur at the left sternal border is almost always audible (in 90–95% of patients), and in roughly half of patients, physical signs of congestive heart failure, including rales, peripheral edema, ascites, and hepatomegaly, are evident at presentation. The earliest report of the use of aortography to diagnose an unruptured aneurysm was published by Falholt and Thomsen in 1953 [16].

Unruptured ASVs that are producing hemodynamic derangements should be repaired. When a congenital ASV has ruptured or is associated with a VSD, prompt surgery is advisable. In patients not surgically treated, the mean survival time after diagnosis of a ruptured ASV has been reported to be 3.9 years, although, if two unusual patients who survived for 10 and 15 years are excluded from this series of 45 cases, the mean survival time would be roughly 1 year [17]. Although death after rupture of an ASV is usually the result of congestive heart failure, patients with this condition have a particular tendency to develop bacterial endocarditis.

Our case was a 55-year-old Asian female who suffered from exertional dyspnea and cardiac murmur. Ruptured ASV to the right atrium without VSD was confirmed by echocardiography and angiography. According to the reported incidence of the various sites of rupture of ASVs, rupture into the right atrium is uncommon in Asians. The prevalence of such rupture in Asian and non-Asian populations is 13% and 35%, respectively [5]. In addition, the basic abnormality in Asians is located leftward and toward the commissural area between the right and left coronary cusps, and the prevalence of rupture into the right ventricle rather than into the right atrium is higher (94% vs 77%).

ASV may be difficult to diagnose clinically but can easily be confirmed by echocardiography. Rupture of an ASV usually results in a large shunt from the aorta to the receiving heart chambers and symptoms of acute heart failure. Physical examination with a harsh continuous murmur and bounding arterial pulses provide a clue to aid differential diagnosis. Surgical correction is indicated after confirmation by echocardiography and cardiac catheterization, if needed. If treatment is initiated as early as possible, the development of congestive heart failure and infective endocarditis will decrease.

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無伴隨心室中隔缺損的主動脈竇動脈瘤破裂到右心房：病例報告及文獻回顧

李碩粲[1] 林宗憲[1] 蘇河名[1] 邱肇基[2] 溫文才[1] 賴文德[1] 許勝雄[1] 林永哲[2]

[1]高雄醫學大學附設中和紀念醫院 心臟血管內科 [2]心臟血管外科

主動脈竇動脈瘤通常伴隨心室中隔缺損。其可能為先天或後天得到的罕見心臟病。主動脈竇動脈瘤少發生在非冠狀動脈竇。一旦主動脈竇動脈瘤破裂通常會產生嚴重的血行動力學變化。若不開刀治療通常預後不良。我們報導一位 55 歲女性病人，其主述為活動時有氣促情形。胸前超音波與主動脈攝影顯示主動脈竇動脈瘤在非冠狀動脈竇，無伴隨心室中隔缺損且動脈瘤破裂到右心房。由於有大量的左到右分流，病人接受手術治療將主動脈竇動脈瘤破裂修補。這個病例呈現一位少見非冠狀動脈竇的主動脈竇動脈瘤破裂到右心房且無伴隨心室中隔缺損。

關鍵詞：動脈瘤，主動脈竇，破裂，右心房，非冠狀動脈竇

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通訊作者：林宗憲醫師

高雄醫學大學附設中和紀念醫院心臟血管內科

高雄市三民區十全一路 **100** 號